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The Relationship of Mirizzi Syndrome and Cholecystoenteric Fistula: Validation of a Modified Classification

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The article by Beltran et al. in this issue of the *Journal* [1] describes the “natural history of Mirizzi syndrome,” including late complications, such as cholecysto-enteric fistula. In western countries, Mirizzi syndrome and cholecysto-enteric fistula are rare conditions because the diagnosis and treatment of symptomatic gallstone diseases are usually made at an earlier stage [2]. Therefore, late complications of chronic inflammation of the gallbladder might be easily undetected, and consequently, inadequately treated due to the lack of experience with these life-threatening conditions. In contrast, in areas with a high incidence of gallstone disease, such as Chile, a significant number of patients who require surgery are referred to the hospital at an advanced stage of chronic inflammation, including Mirizzi syndrome or cholecysto-enteric fistulas.

To better define prognosis and treatment of patients with Mirizzi syndrome, Csendes et al. [3] have proposed a classification consisting of four different types: type I, in which an impacted gallstone leads to an extrinsic compression of the common bile duct; type II corresponds to a cholecysto-biliary fistula secondary to an eroded gallstone involving one-third of the circumference of the common bile duct; type III the fistula involves two-thirds of the circumference; and type IV the fistula involves the whole circumference (Fig. 1). Recently, Csendes et al. [4] added a fifth type to his original classification covering any kind of Mirizzi syndrome in association with a cholecysto-enteric fistula.

In the current article, the same group validated the modified Csendes classification by analyzing a large database of 5,673 patients who underwent cholecystectomy for symptomatic gallstone disease. They demonstrated a strong association between Mirizzi syndrome and the presence of cholecysto-enteric fistula. In addition, the authors showed that the postoperative course strongly depends on the appropriate workup before surgery and the availability of a surgeon experienced in hepatobiliary surgery. We were invited to recapitulate the essentials of the management of Mirizzi syndrome.

The preoperative diagnosis of Mirizzi syndrome is difficult because the clinical and laboratory presentations are unspecific. They include acute abdominal pain, jaundice, and laboratory findings that are consistent with other extrahepatic cholestatic liver diseases. Although ultrasonography (US) remains the “gold standard” to diagnose gallstone disease, an analysis of 16 cases of Mirizzi’s syndrome during a 12 year-period showed that ultrasound was unable to secure the diagnosis in most cases [5]. In a review on this topic, Lai and Lau [6] favored the use of computed tomography (CT) in all suspected cases of a Mirizzi’s syndrome, mostly to rule out a malignancy and to better define the local inflammatory changes. However, the specificity of CT findings to ascertain the diagnosis of Mirizzi’s syndrome is poor, and therefore many would add a more accurate test to visualize the biliary system, such as endoscopic retrograde cholangiopancreatography (ERCP), percutaneous transhepatic cholangiography (PTC), or magnetic resonance cholangiopancreatography (MRCP). In our center, we omit the CT and routinely perform MRCP. The currently reported rate of Mirizzi syndrome correctly diagnosed preoperatively ranges from 10–60% [6].

Surgery is the only curative therapy for a Mirizzi syndrome, because the gallbladder must be removed to treat the

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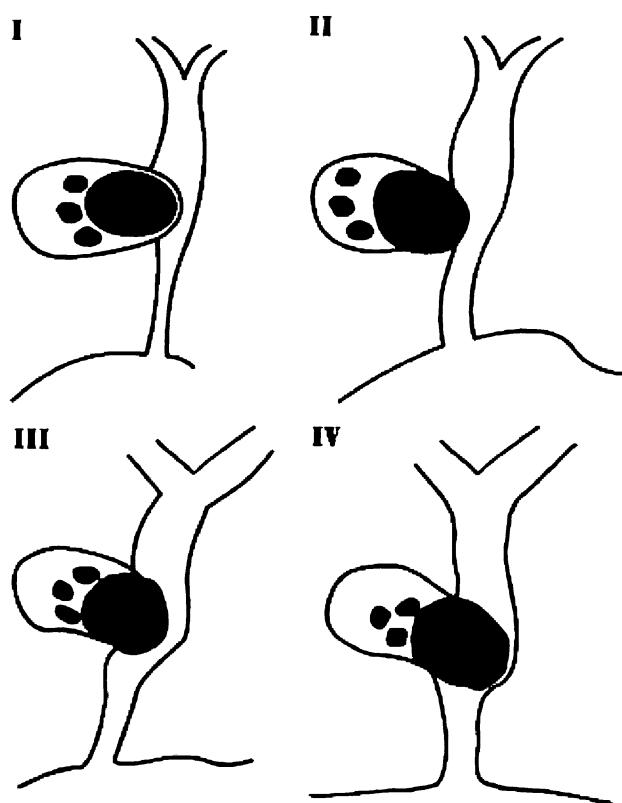


Fig. 1 Schematic representation of Csendes classification for Mirizzi syndrome

underlying disease. If the diagnosis is made preoperatively, particularly in the presence of jaundice, most surgeons will opt for an open approach. However, the diagnosis often is made only during laparoscopic cholecystectomy. Schäfer et al. [7] investigated the outcome of 39 patients with Mirizzi syndrome primarily operated through a laparoscopic approach between 1995 and 1999 in Switzerland. Although the diagnosis was suspected preoperatively in half of the cases, the type of the Mirizzi syndrome could be established only intraoperatively. Due to the inflammatory status, three-quarters of the laparoscopic procedures were converted to an open approach. Twenty-three patients (59%) were treated with cholecystectomy only, whereas additional open bile duct exploration with T-tube insertion

was performed in 13 patients (33%). In three patients (8%), a hepatico-jejunostomy was performed. The reported morbidity rate was <10% with no mortality. The authors recommend a dissection from the fundus of the gallbladder toward Hartmann's pouch, and in presence of severe inflammation, to limit the surgery to a subtotal removal of the gallbladder. In cases of circumferential defects or cholecysto-choledochal fistula, a hepatico-jejunostomy was recommended, whereas placement of a T-tube drain is only sufficient in minor defects of the common bile duct.

Most of the surgeons in the western world have limited experience with Mirizzi syndrome, particularly when the disease is complicated by a cholecysto-enteric fistula. Therefore, a valuable classification system as presented by Csendes provides helpful information for the decision that has to be taken in selecting the surgical therapy of the Mirizzi syndrome. The single most significant factor to minimize postoperative complications relies on the availability of a surgeon experienced in hepatobiliary surgery.

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